Communities Living with Sickle Cell Disease/Trait: Nursing Roles & Collaboration

Robin R. Leger, RN, MS, PhD
University of Connecticut Health Center
rleger@uchc.edu
Overview & Objectives

1) Pathophysiology and management needs of individuals with SCD/T over the lifespan

2) Examples of Nursing Roles & Collaboration with families, providers, & communities:
   - Educating Clinicians & Community
   - Community trait screening
   - Appraising aspects of stigma that may impair access to & quality of care/services

3) Lessons Learned & Future Needs

4) Resources & References
Sickle Cell Disease

- Most common hereditary condition worldwide
- 2 altered hemoglobin genes
- Effects @ 1 in 475; 100,000 in the US
- Different genotypes (SS, SC, SD, etc) and phenotypes (mild, moderate, or severe symptoms & clinical outcomes affecting HRQOL)
- Chronic anemia, hemolysis
- Sickling & inflammation episodes lead to organ damage and disability
SCD in Connecticut

- CT Department of Public Health
  - Newborn screening program
  - Stakeholders Group coordinated by Hospital for Special Care
  - Grant funded initiatives & Public Awareness

- ~ @ 400 Children & 300 Adults with SCD

- ~ 10,000 individuals with Sickle Cell Trait

- Pediatric Sickle Cell Treatment Centers
  - Connecticut Children’s Medical Center
  - Yale New Haven Hospital
Worldwide Distribution

- SCD is found in Africans, Turks, Greeks, Saudi Arabians, Egyptians, Iranians, Italians, Latin Americans and Asiatic Indians.

- SCD is present in one out of four hundred African Americans in the United States. It is the most common genetic disease in this country.

![Worldwide Distribution of Hemoglobin Variants](image)
Hemoglobinopathies a brief overview: Genetics

- Inherited ~ single gene
- Substitution of amino acid
- Pedigree
Sickle Cell Disease

- An inherited, autosomal recessive, disease of red blood cells and endothelium; Single point mutation – valine for glutamic acid.

- Genetic screening ~ many hemoglobinopathy variants
  “SS” is more severe

- Affects the oxygen carrying protein, hemoglobin, in red blood cells (old term: sickle cell anemia). Sickle-shaped red cells interrupt blood flow by blocking small blood vessels (vaso-occlusion event)

- Tissue (at any organ site) that has no blood flow is damaged and causes pain. High risk for disability and organ disease over the life-span
Disease Complications

- Sickle cells become trapped and destroyed in the spleen causing Splenic Sequestration
- Anemia - hemolysis
- Pain episodes
- Hand foot syndrome-Dactylitis, < 2yrs
- Gall Stones
- Strokes (silent) or aneurysms
- Chronic renal disease/failure
- Pneumonia or Acute Chest Syndrome
- Increased Infections
- Bone/joint infarctions/avascular necrosis
- Priapism, > 24hrs dysfunction
- Retinopathy, hearing loss
- Iron Overload from Transfusion Therapy
Dependence ~ Tolerance ~ Addiction

- **Pain Crisis (VOEs):** per year / per individual
  - 90% have 0 – 3/yr
  - 5% have 3 – 12/yr
  - 5% > 13/yr (consume 50% resources)

- **Physical Dependence**
  - Anyone after 7 days continuous opiate use

- **Tolerance**
  - Anyone on continuous opiate- increased dosing is needed

- **Addiction:** Life revolves around drug = @ 5%

- **Pseudo-addiction cycle:** Under treatment → return → under treatment → return = ED & Hospital recidivism!
Managing SCD

- National Newborn Screening - since 1990 in CT
- Prophylactic Penicillin, start age 2 months
- Vaccination Prevnar – pneumococcal
- Pain Management – acute & chronic; life-span
- Prevention of infections
- Chronic blood transfusions
- Antibiotics/Surgery
- Bone marrow transplantation
- Hydroxyurea
- Nitric Oxide
- Increased Fluid Intake
- Healthy lifestyle -> HRQOL!
Sickle Cell Trait

- Sickle cell trait is present in 1/10 African Americans and % varies in others. About half of the hemoglobin in the red cell is sickle and the RBCs will exhibit sickling when under severe conditions of low oxygenation.
- Resistance to Malaria
- Hematuria or blood in the urine is the most common problem.
- Those with trait should be advised of risks of extreme physical activity, severe pressure changes, deep sea diving, dehydration, and possibilities of hematuria -- sever (rhabdomyolysis) & life threatening conditions.
Disparities in Health & Community Services:
Unequal treatment, access, or opportunities due to:
Historical & Environmental Contexts

- Social Economic Status (SES-ism)
- Race
- Ethnic or language (cultural barriers)
- Gender (sexism) * Age (ageism)
- Infectious/contagious/“contaminated”; “it’s in the blood” (L. Pauling)
- Chronic illness
- Mental health condition
- Disability
Hemoglobinopathy Community Testing: Troubled History Current Practice

- Problems with testing included equipment sensitivity (false negatives)
- Discriminatory practices by employers, the military and insurers
- Misinformation
- Stigmatization of the condition; both Disease & Trait Status
- Impaired Access
CT Hemoglobinopathy Educator & Counseling Course: (HRSA 05-027) (DPH 209-0911)

- **Objectives:**
  - Aligning with Newborn Screening Follow-Up
  - Expansion needed for community screening
  - CBO, Clinican and Health Educator Training

- Adapted Cincinnati Intermediate level
- Utilizes CT Clinical Faculty & CT DPH
- SC CBOs as admin. Support & faculty
- Outreach statewide through stakeholders
- National & International Participants
- 4 Levels of Certification
“Making a Family”: High school students demonstrate genetic inheritance with SCD dice.
Hemoglobinopathy Curriculum:

- Overview of genetic patterns of inheritance
- Clinical management of hemoglobinopathies
- Affects on consumers & families across the life-span
- Identification of national guidelines, “best practice” standards of care, & research
- Cultural competencies, legal issues, & counseling skills.
4 Levels of Certification: Based on roles and educational preparation

- Hemoglobinopathy Peer Educator
- Hemoglobinopathy Community Educator
- Hemoglobinopathy Professional Educator
- Hemoglobinopathy Counselor
Mission Statement
To ensure available and accessible quality and comprehensive medical care and support services for children and adults in Connecticut with Sickle Cell Disease and related disorders.

www.cqsc.org

NOW WE HAVE A VOICE!
Sickle Cell Trait: Why do community testing?

- Hemoglobin A ~> altered AS, AC, AE, etc.
- 1 in 10 African Americans + others worldwide
- Diagnosis by blood test: electrophoresis
- Under severe conditions may experience symptoms:
  - Blood in the urine
  - Enlarged spleen
  - Pain episodes or chronic
  - Sudden death?
...taking it to communities
A pilot study was conducted to test for face validity and preliminary psychometrics of two self-reported scales for stigma (surveys for adults living with SCD & family members/caregivers).

Approved by UCHC, IRB # 06-210.

<table>
<thead>
<tr>
<th>Component: Factor Analysis (N = 42)</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Societal impact regarding the disease and isolation</td>
<td>.887</td>
<td>-.328</td>
<td>.146</td>
<td>.291</td>
</tr>
<tr>
<td>2 Personal feelings of shame, rejection, guilt, etc.</td>
<td>.249</td>
<td>.839</td>
<td>-.332</td>
<td>.352</td>
</tr>
<tr>
<td>3 Treatment when in pain &amp; concerns for the future</td>
<td>.163</td>
<td>.433</td>
<td>.789</td>
<td>-.404</td>
</tr>
<tr>
<td>4 Sense of burden and needing assistance</td>
<td>-.354</td>
<td>-.032</td>
<td>.496</td>
<td>.792</td>
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Keep in mind!

- Genetics information is complex and may not be well understood by communities and families; results may raise questions about family histories.

- Communities have unique attributes which can make providing awareness and health services both challenging & rewarding.

- Community-Based Participatory Research is both a method & a guiding principle for practice.
### Living with Sickle Cell Disease:

**“Old School” vs. New “Improved” Model**

<table>
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<tr>
<th>Hematology Specialty Clinic &amp; Crisis Management via ED</th>
<th>Chronic Illness Model (Wagner): Collaborative and Continuous across environments</th>
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<tbody>
<tr>
<td>Pediatric Care</td>
<td>Life-span Approach</td>
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<tr>
<td>Regional Tertiary Medical Centers</td>
<td>Community Health Centers &quot;Medical Homes&quot;</td>
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<tr>
<td>Paternalistic Approach</td>
<td>CBPR with Community-Based Organizations (CBOs)</td>
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<tr>
<td>Prevention of VOE &amp; Decrease Pain</td>
<td>Preventing organ damage &amp; disabilities</td>
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Major Areas for Intervention

- Improved Access to Care in Racial & Ethnic Disparities
- Improved Quality of Care for Acute on Chronic Conditions
- Continuity Across Environments of Care
- Pain Management (Acute & Chronic)
- Prevent Secondary Conditions!
- Functional Status (Ed., Voc., Psycho-social, Rehab)
- Schools & Athletics
- Health-Related Quality of Life
- Equity in Allocations of Resources and Research
Resources

- **211 Info Line**
  - The Genetic Alliance [www.geneticalliance.org](http://www.geneticalliance.org)
  - Citizens for Quality Sickle Cell Care (CQSCC)
    100 Arch St. New Britain, CT 06050 [www.cqscc.org](http://www.cqscc.org)
    860-223-7222 [citizensforquality@sbcglobal.net](mailto:citizensforquality@sbcglobal.net)
  - Sickle Cell Disease Association of America of Southern Connecticut
    189 State St. Bridgeport, CT 06604
    203-366-8710 [scdaasoutherncnt@sbcglobal.net](mailto:scdaasoutherncnt@sbcglobal.net)
  - Sickle Cell Disease Association of America, Inc.
    231 East Baltimore Street, STE 800
    Baltimore, MD 21202 [scdaa@sicklecelldisease.org](mailto:scdaa@sicklecelldisease.org)
  - Grady Health System; Sickle Cell Information Center
    [www.SCInfo.org](http://www.SCInfo.org)
Select References


